Multiple neuroendocrine tumors in the stomach, duodenum and pancreas of a MEN1 patient

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Dear Editor,

A 67-year-old female patient came to our hospital complaining of recurrent abdominal pain and diarrhea in the previous ten years. She underwent gastroscopy three years previously, which revealed multiple polyps in the stomach and submucosal tumors in the pylorus (Fig. 1A) and duodenal bulb (Fig. 1D). Endoscopic submucosal dissection (ESD) was performed for all lesions at that time, and a pathological diagnosis of G1 neuroendocrine tumor (NET) was established (Fig. 1B, C, E and F). However, her symptoms recurred half a year before, and laboratory evaluation showed an elevated level of serum gastrin (1,000 pg/ml). Contrast-enhanced abdominal computed tomography (CT) scan demonstrated enhancement in the neck of the pancreas (Fig. 1G). No abnormalities were observed via thyroid ultrasound or cranial magnetic resonance (MR), but superficial gastritis was identified by upper endoscopy. Endoscopic ultrasound (EUS) displayed a hypoechoic lesion with a uniform echo in the neck of the pancreas, which was contrast-enhanced (Fig. 1H). Pancreatic tissues were collected through EUS-guided fine needle aspiration (EUS-FNA), and histopathologic findings revealed G1 NET. Thereafter, the patient received laparoscopic enucleation of the pancreatic tumor. Postoperative pathological examination of the resected
specimen confirmed a gastrin-producing G1 NET in the pancreas (Fig. 1I and J), which was homologic to previous lesions, and one metastasis to peripancreatic lymph node. Her serum gastrin level decreased to 270.00 pg/ml after surgery.

Discussion
Gastroenteropancreatic neoplasms (GEP-NENs) often arise from the pancreas (31.5 %), rectum (29.6 %), stomach body (15.4 %) and cardia (11.6 %) (1). Gastrinoma, a common type of GEP-NENs, is often sporadic, rarely manifesting as multiple endocrine neoplasia type 1 (MEN1) (2). According to the National Comprehensive Cancer Network (NCCN) guidelines, MEN1 is clinically diagnosed when two or more of the followings are present: primary hyperparathyroidism, pituitary adenoma, duodenal NET, pancreatic NET, or foregut NET (bronchial, thymic, or gastric) (3). In this case, the middle-aged female patient was found to have multiple gastrinomas in the pylorus, duodenum and pancreas, together with metastases in the peripancreatic lymph node. Thus, we established a clinical diagnosis of MEN1-related gastrinomas (gastric-NET/duodenal-NET/pancreatic-NET, G1) with lymph node metastases. It is reported that MEN1 is closely related to MEN1 gene mutations (4). Regular follow-up is recommended for MEN1-associated pancreatic gastrinomas less than 2 cm in diameter, and surgical resection for those larger than 2 cm. In the case of liver metastasis, options may rely on surgery, radiofrequency ablation, hepatic artery embolization, or targeted molecular drugs, such as sunitinib. Regular follow-up with serum gastrin levels and 68Ga-Dotatate PET/CT should be used to detect recurrences and metastases in a timely manner (2).

References